



**SERVIZIO SANITARIO REGIONALE  
EMILIA - ROMAGNA**  
Istituto Ortopedico Rizzoli di Bologna  
Istituto di Ricovero e Cura a Carattere Scientifico



## “Osteofibrous Dysplasia (Kempson-Campanacci’s disease): Long Term Follow-up Study on Natural History, Results of Treatment and Relationship with Adamantinoma”

<b><u>Study code</u></b>	<b>DOFOL</b>
<b>Sponsor’s Name and Address:</b>	Istituto Ortopedico Rizzoli Via di Barbiano 1/10 40136 Bologna Italy
<b>Study Number/Version/Date:</b>	Vers 1.0 08 Mar 2019
<b>Coordinating Center:</b>	IRCCS Istituto Ortopedico Rizzoli Pathology Unit Via Pupilli 1 40136 Bologna, Italy
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<b>Scientific/Medical study responsible and developer</b>	Alberto Righi MD PhD
<b>Methodology:</b>	Retrospective study (Single institution case series review of clinical data)
<b>Type:</b>	Academic
<b>Founding:</b>	None
<b>Principal Investigator Signature</b>	I confirm that I’ve read this protocol and I accept to run the study in compliance with what is stated in the protocol and with the ICh-GCP and all applicable law  Alberto Righi MD  Firma  _____

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## BACKGROUND

Osteofibrous dysplasia, also called Kempson-Campanacci disease, is a rare benign fibro-osseous lesion typically involving the tibia or the tibia and fibula, usually diagnosed during early childhood or pre-adolescent age (1,2). Despite benign self-limiting course it may cause anterior bowing, bone expansion and moderate pain. Osteofibrous dysplasia is rare, with reported incidence of 0.2% of all primary bone tumors (1-3). It is slightly prevalent in males and is usually diagnosed within the first 2 decades of life, most commonly in children less than 12 years old (1-4). Since malignant progression to adamantinoma has been reported, management of this condition remains somewhat controversial (1-7). The potential role of osteofibrous dysplasia as precursor lesion capable of progression to adamantinoma has received support by the more recent description of osteofibrous dysplasia-like adamantinoma or differentiated adamantinoma, an intermediate lesion possibly representing the missing link between osteofibrous dysplasia and adamantinoma (5-7). However, consensus has not been reached on the histologic criteria differentiating osteofibrous dysplasia, osteofibrous dysplasia-like adamantinoma and adamantinoma and progression to adamantinoma has not been convincingly documented so far (1-7).

## OBJECTIVE OF THE STUDY

The aim of the present study is to obtain long term follow-up in patients with osteofibrous dysplasia, to assess natural history of the disease, late results of treatment and in particular the potential and risk of progression to adamantinoma.

The study will exam all the cases of osteofibrous dysplasia treated between 1943 and 2018.

## STUDY DESIGN

This is single institution cases series review of histological and clinical data

## POPULATION

### Inclusion criteria

- 1) Male and female patients treated at Rizzoli Institute from 01 Jan 1943 to 31 Dic 2011
- 2) Diagnosis of osteofibrous dysplasia pathologically confirmed
- 3) Written informed consent prior to any study-specific analysis and/or data collection

According to the Italian *“Autorizzazione generale n. 9/2016 al trattamento dei dati personali effettuato per scopi di ricerca scientifica”* of the Privacy Tutor (and the corresponding regulation in the other participating countries) the informed consent is not required to be obtained by the

deceased subjects, as long as all the other enrolment criteria are met and the study has been approved by the Ethic Committee (refers to INFORMED CONSENT section).

## **MATERIAL AND METHODS**

We will retrieve from the database of the Rizzoli institute all the cases with a histological diagnosis of Ewing sarcoma from 01 Jan 1943 to 31 Dic 2011.

We expect to find approximately 55 cases.

We will review all the medical records, radiological imaging, and histological data of these cases.

## **STATISTICS**

To the case series will be applied a descriptive statistic.

## **ENROLLMENT PROCEDURE**

Patients considered eligible will be included in the study, after providing a written informed consent.

Since we will include cases of several decades ago, it would be possible that some eligible subjects will be deceased.

## **DATA COLLECTION**

Clinical data will be retrieved by patient charts.

A protocol-specific CRF reporting the results of the review will be provided.

A CRF is required and should be completed for each included subject.

## **ETHICS AND QUALITY ASSURANCE**

The clinical trial protocol and its documents will be sent before initiating the study to the competent Authorities and Ethics Committees of each participating country for its approval.

The responsible investigator will ensure that this study is conducted in agreement with either the most updated Declaration of Helsinki and all the international and local laws that apply to clinical trials and to patient protection.

The protocol has been written, and the study will be conducted according to the principles of the ICH Harmonized Tripartite Guideline for Good Clinical Practice (ref: <http://www.emea.eu.int/pdfs/human/ich/013595en.pdf>).

**INFORMED CONSENT**

All patients will be informed, by the investigator, of the aims of the study, the possible risks and benefits that will derive from the study participation. \*

The Investigator must clearly inform that the patient is free to refuse participation in the study and that can withdraw consent at any time and for any reason.

They will be informed as to the strict confidentiality of their patient data, but that their medical records may be reviewed for trial purposes by authorized individuals other than their treating physician.

The informed consent procedure must conform to the ICH guidelines on Good Clinical Practice. This implies that "the written informed consent form should be signed and personally dated by the patient or by the patient's legally acceptable representative".

The Investigator must also sign the Informed Consent form, and will keep the original at the site and a copy of the original must be handed to the patient.

The competent ethics committee for each Institution participating to the study must validate local informed consent documents before the study can be opened. It will be emphasized that the participation is voluntary and that the patient is allowed to refuse further participation in the study whenever he/she wants. This will not prejudice the patient's subsequent care.

Due to the high incidence of mortality of the disease under investigation, it would be possible that some potential eligible subjects will be deceased.

In order to allow and promote the increase in the knowledge of this rare disease that could be beneficial for other patient that are or will be affected, according to the Italian *"Autorizzazione generale n. 9/2016 al trattamento dei dati personali effettuato per scopi di ricerca scientifica"* of the Privacy Tutor and the corresponding regulation in the other participating country, as well as the EU General Data Protection Regulation 679/2016 (that will be applicable from 25 May 2018), the informed consent is not required to be obtained by the deceased subjects according the aforementioned laws/dispositions.

**CONFIDENTIALITY**

In order to ensure confidentiality of clinical trial data as disposed the national and European applicable regulation, data will be only accessible for the trial Sponsor and its designees, for

monitoring/auditing procedures, the Investigator and collaborators, the Ethics Committee of each corresponding site and the Health Authority.

Investigator and the Institution will allow access to data and source documentation for monitoring, auditing, Ethic Committee revision and inspections of Health Authority, but maintaining at all times subject personal data confidentiality as specified in the “Directive 95/46/EC of the European Parliament and of the Council of 24 October 1995”.

The Investigator must guarantee that patient anonymity is kept at all times and their identity must be protected from unauthorized persons and institutions.

All patients included in the study will be identified with a numeric code, so that no identifiable personal data will be collected (pseudo anonymization)

The Investigator must have and conserve a patients’ inclusion registry where it figures the personal data of the patient: name, surname, address and corresponding identification code into the study, this register will be kept on the Investigator File.

#### **PUBLICATION OF RESULTS**

The results from this study will be published or shown at scientific conferences.

The final publication of the study results will be written by the Principal Investigator.

#### **SPONSOR ROLE AND RESPONSIBILITY**

The sponsor is the sole owner of the data and is responsible of all the clinical trial activities from study design, development, data collection, management, analysis, interpretation of data, writing and the decision to submit the report for publication written by the Principal Investigator,

**REFERENCES**

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